



# Papillary Carcinoma in Thyroglossal Duct Cyst and Thyroid Gland with Hashimoto

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## Abstract

**BACKGROUND:** The ectopic thyroid gland is a rare condition. In most cases, ectopic thyroid tissue is the only functioning thyroid tissue, but it may rarely be associated with a normal thyroid gland. Ectopic thyroid carcinoma within a thyroglossal duct cyst is a very rare entity, with controversial individual approaches.

**CASE PRESENTATION:** This encouraged us to present the case of a 31-year-old woman, with papillary thyroid carcinoma in the thyroid gland and in the thyroid ectopic tissue within the thyroglossal duct. After suspicion of thyroid malignancy in the ultrasonography of the neck, FNA-biopsy was performed on the thyroid nodule and the thyroglossal duct cyst.

**CONCLUSION:** Since the pathological exam revealed a papillary carcinoma in the eutopic and ectopic thyroid tissues, the Sistrunk procedure and total thyroidectomy was done, followed by radioiodine ablation.

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## Introduction

Ectopic thyroid tissue is a rare occurrence with a prevalence of about 1 per 100.000–300.000 people [1], [2], [3]. The most common anatomic classification of thyroid ectopia is the lingual subtype [4], [5]. Sublingual thyroids, that are much less common, may be found along the course of the thyroglossal duct at either supra or infrahyoid locations [4], [5]. Interestingly, females represent their majority, more often at the age of puberty, pregnancy, and menopause [4], [6]. Depending on the location of ectopic tissue, patients may refer to dysphagia, dysphonia, and/or dyspnea [3], [6].

The majority of thyroglossal duct cysts (TGDCs) are presented as an anterior, painless, infrahyoid mass. Most of them are evident from incidental palpation of the neck during a physical examination by the pediatrician [7], [8]. Ectopic thyroid tissue within TGDCs is a common finding, although only in about 1–6.5% of these cases become malignant and particularly develops papillary thyroid carcinomas [5], [9], [10]. In most cases, thyroid carcinoma is diagnosed incidentally by histological investigation after resection of a TGDC. In cases where

ectopic thyroid is present, and cancer cells are evident in both eutopic and ectopic thyroid tissues, it is difficult to determine the true origin of thyroid carcinoma [11]. The diagnostic steps involve ultrasonography and/or magnetic resonance tomography of the neck, followed by FNA biopsy [12]. Therapy of choice in such cases is the surgical resection of TGDCs by Sistrunk procedure and total thyroidectomy [6], [7], [8].

## Case Presentation

### Consent for publication: Footnotes

A 31-year-old woman presented at our clinic with a 2 months history of a slowly enlarging mass located in the anterior midline of her neck. She referred to dysphagia and voice changes. A physical exam revealed a soft, painless, and mobile mass in the submandibular region, and another palpable mass, with limited mobility in the right inferior part of the thyroid gland (Figure 1a and b).

An ultrasound examination revealed a supra laryngeal cyst of 21.7 mm in diameter cranial to the



Figure 1: Clinical photos. (a) Preoperative photo, (b) Postoperative photo

hyoid bone. It also identified changes in the structure of the thyroid gland (Figure 2a and b).

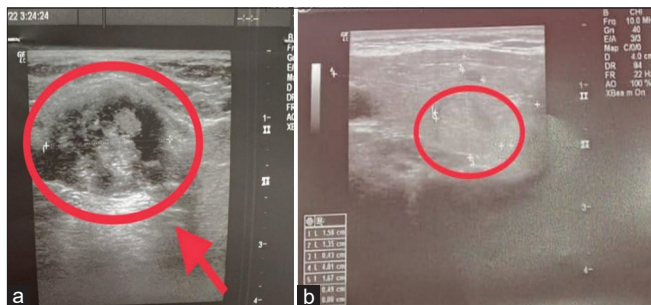


Figure 2: Ultrasound of the neck. (a) The thyroglossal duct cyst. (b) The thyroid nodule

In the cervical anterior median line region, in the middle 1/3, a cystic lesion up to 21.7 mm is seen, with irregular echogenic vegetation in it, with vascularization after color Doppler. The thyroid structure was heterogeneous, with multiple hypoechoic and anechoic areas. In the inferior part of the thyroid right lobe is seen a heterogeneous nodule 15.8 mm, with necrotic areas and microcalcifications inside it, with high intranodular and perinodular vascularization. No cervical lymph nodes were detected.

FNA biopsy was performed in both suspicious areas: The thyroid nodule and the thyroglossal duct cyst. In both areas, elements of papillary carcinoma were identified (Figure 3).

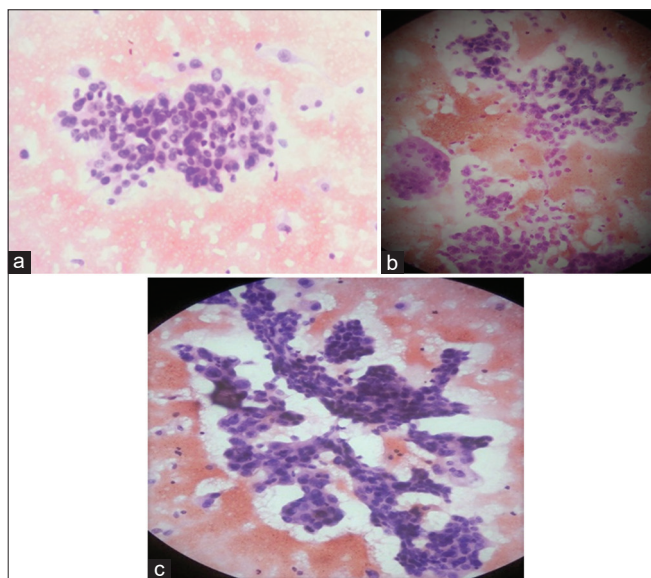


Figure 3: (a-c) Pre-operative FNA biopsy images

Cytologic features in FNA-biopsy: Cleared nuclei, overlapping nuclei, nuclear grooves, and intranuclear inclusions. Such cellular characteristics are classified as Bethesda 5-6.

The patient underwent specific thyroid function blood tests, from which the thyroid showed normal function [FT3 – 33.0 (23–62 pmol/l), FT4–12.8 (10.6–19.4 pmol/l), TSH – 4.7 (0.3–5.0 μIU/ml)], and positive antithyroid antibodies [AntiTPO – 140.1 (< 8.0 IU/ml), AntiTg - 72.9 (< 18 IU/ml)]. Thyroglobulin level was 226.3 (3.5–77 ng/ml). The patient had not previously used any medication for the thyroid gland.

**Treatment**

The patient underwent a Sistrunk procedure, total thyroidectomy, and bilateral neck dissection (Figure 4a and b). Intraoperatively, the thyroglossal duct cyst was found separate from the thyroid gland but in the same anatomical plane. On the right part of the neck, there were three enlarged cervical lymph nodes that were resected. Both upper and lower right parathyroid glands were excised and implanted after negative histological results by frozen section biopsy. Both upper and lower left parathyroid glands remained *in situ*. During the intervention, the external branch of the superior laryngeal nerve and recurrent laryngeal nerves was identified bilaterally and preserved.

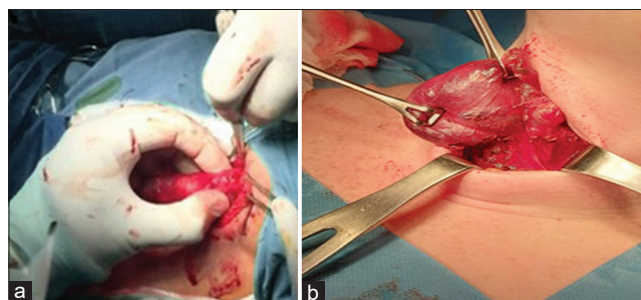


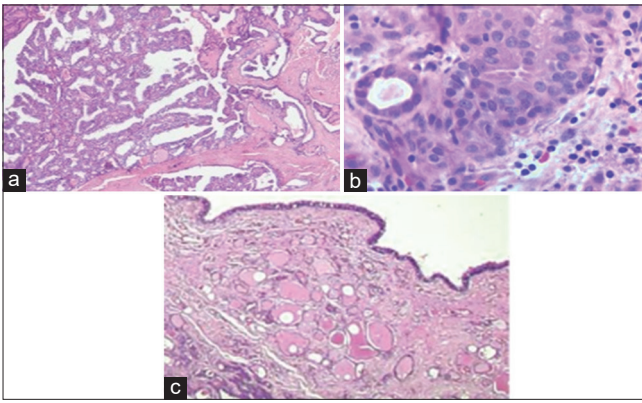
Figure 4: Surgical images. (a) Surgical resection of the thyroglossal duct cyst by the Sistrunk procedure. (b) Surgical resection of the thyroid gland (total thyroidectomy)

The histopathological examination of the removed thyroid tissue revealed papillary thyroid cells (Figure 5).

Histopathological evaluation of the resected lymph nodes revealed a negative result for malignancy. Four weeks after the surgery, the patient was subjected to radioactive iodine therapy (30 MCI). At present, the patient is on TSH suppression therapy, with 100 mcg of Levothyroxine per day. The last thyroglobulin level is 0.38 ng/ml.

**Discussion and Conclusions**

The first case of ectopic thyroid was published in 1869 by Hickman, who described a lingual thyroid in



**Figure 5: Histopathological description.** (a) In this figure, we note the papillary carcinoma, the true papillae with multiple branching with fibrovascular cores. (b) In this figure, the cells are presenting changes in nuclear size and shaped ground glass nuclei with nuclear membrane irregularity. (c) This photo displays the epithelium of the cyst. The lining epithelium is a single layer of flattened cells. Around the cyst are thyroid follicles

a newly-born baby that underwent suffocation 16 h after birth as a consequence of a tissue mass that caused upper airway obstruction [1], [3], [5]. The thyroid gland in embryogenesis develops as the first pharyngeal derivative, primarily appearing as an invagination in the floor of the pharynx. During this process, after migrating caudally to the trachea, it remains attached to the tongue by the thyroglossal duct, which usually undergoes atrophy after birth. If it fails to involute, it can persist as a cyst, a duct, or ectopic tissue, which is localized in the midline between the base of the tongue and the pyramidal lobe of the thyroid gland [1], [3]. Clinically, most thyroglossal duct cysts are benign and present as slow-growing, asymptomatic neck masses [9], [12]. Criteria for TGDC diagnosis were first published by Roses *et al.* [13]. The TGDCs are considered to be caused by mutations in genes such as the thyroid transcription factors thyroid transcription factor 1 (TTF1), thyroid transcription factor 2 (TTF2), and paired box gene 8 (PAX8) responsible for the development of thyroid follicular cells [13], [14]. The initial assessment of all TGDCs should always include consideration of possibly existing ectopic thyroid tissue and TGDCa. Thus, clinical examination, the investigation of the cervical lymph nodes, thyroid functional tests, neck imaging, and thyroglobulin test is necessary [11], [15], [16]. It is important to be investigated the presence or not of thyroid tissue in the thyroid region [1], [2], [4], [5], [6]. In the case of a present thyroid gland, the Sistrunk procedure would remove all functionally active thyroid cells [7]. Papillary thyroid carcinoma is the most common form of TGDCa, followed by squamous cell carcinomas [12], [14], [17], [18]. Their management strategy continues to be indefinite, with controversial opinions about thyroidectomy in cases with normal thyroid morphology on ultrasonography. The decision on how to manage them is easier to take in cases where the ultrasound examination reveals nodule(s) in the thyroid

gland. A positive FNA biopsy for thyroid malignancy in the eutopic thyroid tissue completely clarifies the next step. In all such cases, radioiodine ablation should be evaluated after the Sistrunk procedure and total thyroidectomy [9], [12]. We report a young patient presenting with Hashimoto thyroiditis and papillary thyroid carcinoma in the thyroglossal duct cyst and the thyroid gland. The neck imaging by ultrasound was suspicious for malignancy in the thyroglossal duct cyst and the thyroid nodule too. Mandatory for standardized therapeutic management of these rare carcinomas is the understanding of their origin. In fact, there are two possibilities for the development of a TGDCa: 1 - the cyst is the primary origin of the malignant cells; and 2 - TGDCa is a distant metastasis of a pre-existing thyroid carcinoma within the thyroid gland itself [2], [12], [19]. In our case, the second possibility is more likely. However, whatever the origin, FNA biopsy positive for malignancy helped us to consider both the Sistrunk procedure and total thyroidectomy with subsequent radioiodine ablation as the most reasonable therapeutic approaches. It is an interesting fact that other authors suggest this strategy even in cases where the thyroid gland is normal, without malignant nodules [18], [20]. This is because the carcinoma may be multifocal and there may be a lymphatic invasion of the thyroid gland. Besides this, in their opinion, this is the best strategy to ensure a correct follow-up by thyroglobulin measurement [18]. Based on the available literature for this pathology, the Sistrunk procedure may be enough for a TGDCa under 1 cm. In all other cases, like ours, the possibility of a total thyroidectomy should be considered, followed by radioactive iodine therapy [18], [19], [20], [21], [22]. The progress of our case so far has been satisfactory. Hence, 3 months after the radioactive iodine therapy, our patient underwent the ultrasonography of the neck and the thyroglobulin level measurement. The ultrasonography of the neck did not show any residual thyroid tissue and/or lymph nodes present in this region. The thyroglobulin level was 0.38 ng/ml, on TSH suppression therapy with 100 mcg of Levothyroxine per day. For this case, our follow-up plan includes the ultrasonography of the neck and the thyroglobulin level test every 6 months, for a 5-year period.

## Footnotes

Abbreviations: TGDC = thyroglossal duct cyst, TGDCa = thyroglossal duct cyst carcinoma, FNA biopsy = Fine-Needle Aspiration biopsy, TTF1 = Thyroid Transcription Factor 1, TTF2 = Thyroid Transcription Factor 2, PAX8 = Paired box gene 8.

## Consent for Publication

After being informed about publishing her case, the patient provided her consent, approved by the Ethics Committee of Tirana Medical University (Protocol no. 361/2022).

## Availability of Data and Material

The authors declare that the data supporting the findings of this study are available within the article.

## Authors' Contributions

ThY and HE made the initial diagnosis of the thyroglossal duct cyst and the thyroid nodule. ND and AM performed the histological examination of the thyroglossal duct cyst and thyroid gland. ThY took care of the patient after diagnosis of thyroid carcinoma and managed therapy as well as the postoperative follow-up examinations. SE was the major contributor to writing this manuscript. All authors listed above have participated in critical revision of the article and gave their final approval of this version of the manuscript to be published.

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